

CASE REPORT

Case of adult-onset Coats' disease with epiretinal membrane treated with 25-gauge pars plana vitrectomy

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Abstract : We describe a case of untreated adult-onset Coats' disease with a proliferative epiretinal membrane (ERM) treated successfully with 25-gauge pars plana vitrectomy (25GPPV). A 26-year-old man presented with a 3-week history of decreased vision in his left eye. At the initial examination, the decimal best-corrected visual acuity (BCVA) was 0.7 in the left eye. Ophthalmoscopy revealed the typical appearance of Stage 2A Coats' disease but with a proliferative ERM in the posterior pole. The patient received 2 monthly intravitreal injections of 2.5 mg bevacizumab, 5 laser photocoagulations to the area of telangiectasia, and 1 session of cryoretinopexy. Nine months after the initial visit, a traction by the ERM on the parafoveal area developed causing macular edema which reduced the BCVA to 0.3. He underwent 25GPPV with the removal of the ERM. In addition, the peripheral telangiectasia was treated intraoperatively with both laser photocoagulation and cryoretinopexy. Postoperatively, the traction to the parafoveal area was released and the BCVA improved to 0.6 which remained stable during the follow-up period of 13 months. We conclude that 25GPPV combined with ERM peeling, laser photocoagulation, and cryoretinopexy can be effective for adult-onset Coats' disease associated with an ERM. *J. Med. Invest.* 62 : 85-88, February, 2015

Keywords : Coats' disease, cryoretinopexy, epiretinal membrane, intravitreal bevacizumab, laser photocoagulation, pars plana vitrectomy

INTRODUCTION

Coats' disease was first reported in 1908 (1), and it is a nonhereditary disease characterized by idiopathic retinal telangiectasia, intraretinal and subretinal exudation, and exudative retinal detachment. There is no sign of inflammation. In addition, there may be capillary nonperfusion, aneurysmal formation, and massive lipid deposition (2). Degeneration of the endothelial and mural cells of the retinal arteries and veins has been suggested to be the primary pathologic defect of this disease, and the changes can result in dilation and convolution of the retinal vessels leading to exudation (3). Light and electron microscopic examinations of the retina of younger Coats' disease patients reveal a loss of endothelial cells and pericytes of the affected vessels with mural disorganization and breakdown of the blood-retinal barrier (4). Coats' disease usually occurs unilaterally in young men and, if untreated, can lead to total retinal detachment and secondary glaucoma, sometimes requiring enucleation (5).

Observation is generally recommended in the earlier stages of Coats' disease (5). In cases with a progression to retinal telangiectasia and secondary subretinal exudates, laser photocoagulation and/or cryoretinopexy are recommended. Patients at an advanced stage are managed by pars plana vitrectomy, subretinal fluid drainage, and silicone oil tamponade (6-9). However, the prognosis is usually not good (10).

Coats' disease less commonly presents in later childhood with similar features. If Coats' disease is diagnosed in older children, the condition usually progresses at a slower rate (11). Several studies have shown that the earlier the age of presentation, the more severe will be the progression of the disease and the greater the

likelihood of eventual enucleation (5, 11, 12). Smithen *et al.* (11) reported that the characteristics of older patients with adult-onset Coats' disease included a limited area of involvement, slower progression, and hemorrhages near the larger vascular dilatations. None of the patients progressed to the end-stage of Coats' disease with iris neovascularization and total exudative retinal detachment. An epiretinal membrane (ERM) was present in 2 of the 13 cases adult-onset Coats' disease presented by Smithen *et al.* (11), although the details and the management of the ERM were not presented.

Usually, an ERM develops iatrogenically, e.g., after excess retinal photocoagulation or cryoretinopexy (10, 13). Appiah *et al.* (13) studied 187 consecutive patients with a diagnosis of secondary ERM, and reported that the surgical causes were cataract extraction in 41.7%, scleral buckle in 18.9%, laser photocoagulation in 9.7%, and cryoretinopexy in 8.0%. In the same way, an ERM is detected after laser photocoagulation and/or cryoretinopexy in eyes with Coats' disease. In a case series of 150 patients with juvenile or adult-onset Coats' disease, 2 of 79 eyes (2.5%) with a final decimal visual acuity of < 0.1 had an ERM suggesting that ERM is rare but can cause severe decrease of the visual acuity (5). Rishi *et al.* (14) reported the clinical features, treatment, and outcomes in 307 eyes with Coats' disease. They reported that an ERM was a complication after treatment in 13 of the 307 eyes (4.4%). However, there was no mention of any treatment of the secondary ERM in the above-mentioned 2 large case series (5, 14). Yadav *et al.* (15) reported a case that had vitrectomy for an ERM secondary to treatment of juvenile Coats' disease. In their case, the foveal contour improved and the vision improved from 0.17 to 1.0 after surgery indicating excellent surgical outcome.

In cases of untreated Coats' disease, only several cases of an ERM have been published (3, 10, 11, 16). To date, only two cases of juvenile (3) or adult-onset (16) untreated Coats' disease that underwent vitrectomy for ERM have been published, and the surgical outcomes were good.

We report a case of adult-onset Coats' disease with an ERM

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without prior treatment which was successfully treated with 25-gauge pars plana vitrectomy (25GPPV).

CASE REPORT

A 26-year-old man presented with a 3-week history of reduced vision in his left eye. There was no history of radiation treatment, ocular trauma, inflammation, or systemic disease. At the initial visit, the decimal best-corrected visual acuity (BCVA) was 1.5 in the right eye and 0.7 in the left eye. The intraocular pressure was 18 mmHg in the right eye and 17 mmHg in the left eye. The anterior segment of both eyes and fundus of the right eye were normal. Fundus examination of the left eye showed retinal telangiectasia with retinal and preretinal hemorrhage, subretinal fluid, and yellow lipid exudation in the inferotemporal quadrant indicating Stage 2A (telangiectasia and extrafoveal exudation) Coats' disease (Figure 1a) (5). The proliferative ERM extended from the inferior margin of the optic disc to the inferotemporal mid-periphery. A shallow incomplete posterior vitreous detachment (PVD) was detected. Fluorescein angiography revealed intense leakage from the peripheral telangiectasia (Figure 1b) and aneurysmal dilatations with adjacent capillary non-perfusion. There were no signs of macular telangiectasia. Spectral-domain optical coherence tomography (SD-OCT) showed that the ERM was causing traction on the disc margin (Figure 1c). A diagnosis of stage 2A Coats' disease accompanied by a proliferative ERM was made. The patient had not been treated before our examinations indicating that the ERM was not due to prior procedures.

The patient received 2 monthly intravitreal injections of 2.5 mg bevacizumab, 5 laser photocoagulations to the area of the telangiectasia, and 1 session of cryoretinopexy. Four months after the initial visit, SD-OCT showed an increase in the traction on the disc margin and slight cystic macular edema (Figure 2a-c). The SD-OCT images also showed a flattening of the foveal contour. Inferior dislocation of macula in addition to a flattened foveal contour and cystic macular edema suggested possible macular involvement. However, the BCVA remained 0.7. The external limiting membrane and photoreceptor inner/outer segment junction lines were continuous, although the reflectivities of these lines were slightly reduced.

Nine months after the initial visit, traction on the parafoveal area developed causing moderate cystoid macular edema, and the BCVA decreased to 0.3 (Figure 2d-f). There was an ERM superior to the fovea, which was not seen 4 months after the initial visit, suggesting that the ERM might progress after intravitreal bevacizumab, laser photocoagulation and cryoretinopexy. Two weeks later, he underwent 25GPPV with the removal of the ERM and air tamponade of the retina. Intraoperatively, the peripheral telangiectasia was treated by both laser photocoagulation and cryoretinopexy. No intraocular dye was used to stain the ERM. Intentional internal limiting membrane (ILM) peeling was not performed during the surgery. Thirteen months after the 25GPPV, the traction on the parafoveal area was not present, and the BCVA improved to 0.6 (Figure 2g-i). However, macular edema and fine folds of the retinal surface remained, and a small ERM regrowth developed in the region of the superior arcade vessels. The BCVA remained stable during the 13 months of follow-up after the surgery.

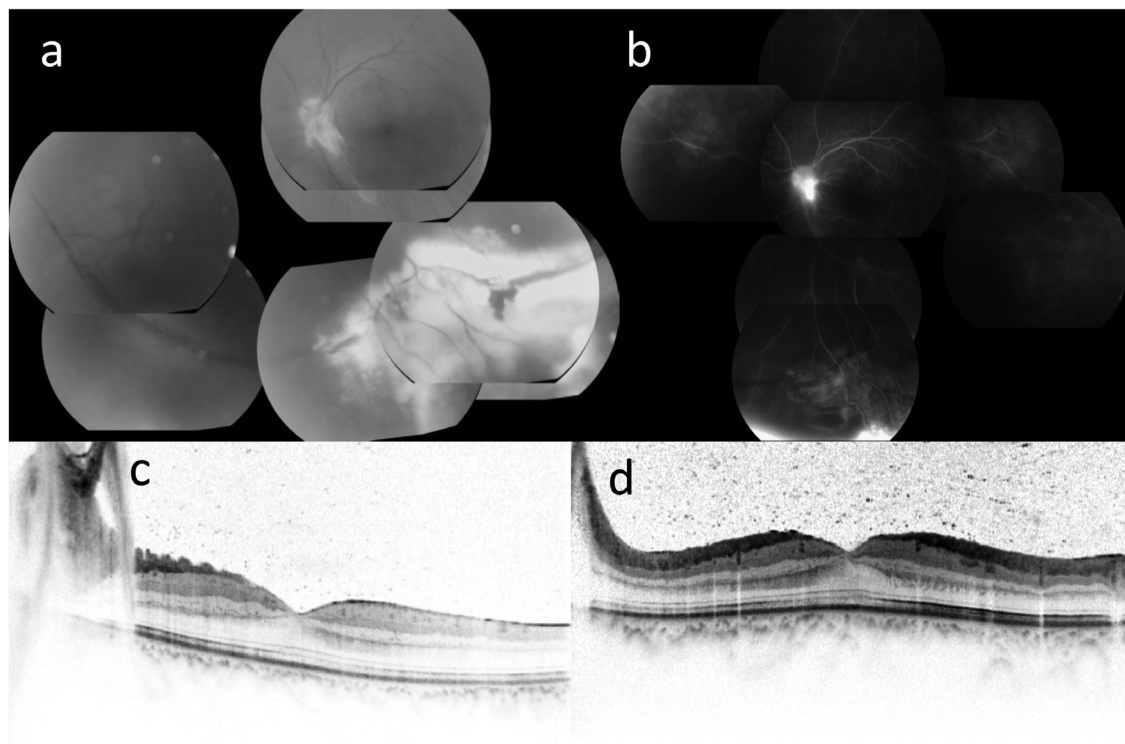


Figure 1. Fundus photograph, fluorescein angiogram, and spectral-domain optical coherence tomographic (SD-OCT) images of an eye with Coats' disease at the initial visit. The best-corrected visual acuity was 0.7.

a : Fundus photograph showing retinal telangiectasia inferotemporally with yellow lipid exudation. Retinal and preretinal hemorrhage maybe along with partial vitreous detachment is observed. An epiretinal membrane (ERM) can be seen extending from the inferior margin of the optic disc to the inferotemporal midperiphery.

b : Fluorescein angiogram showing intense leakage from the peripheral telangiectasia. There were no signs of macular telangiectasia.

c : SD-OCT image of a horizontal scan through the fovea shows an ERM causing traction on the disc margin.

d : SD-OCT image of a vertical scan through the fovea shows an almost normal foveal contour.

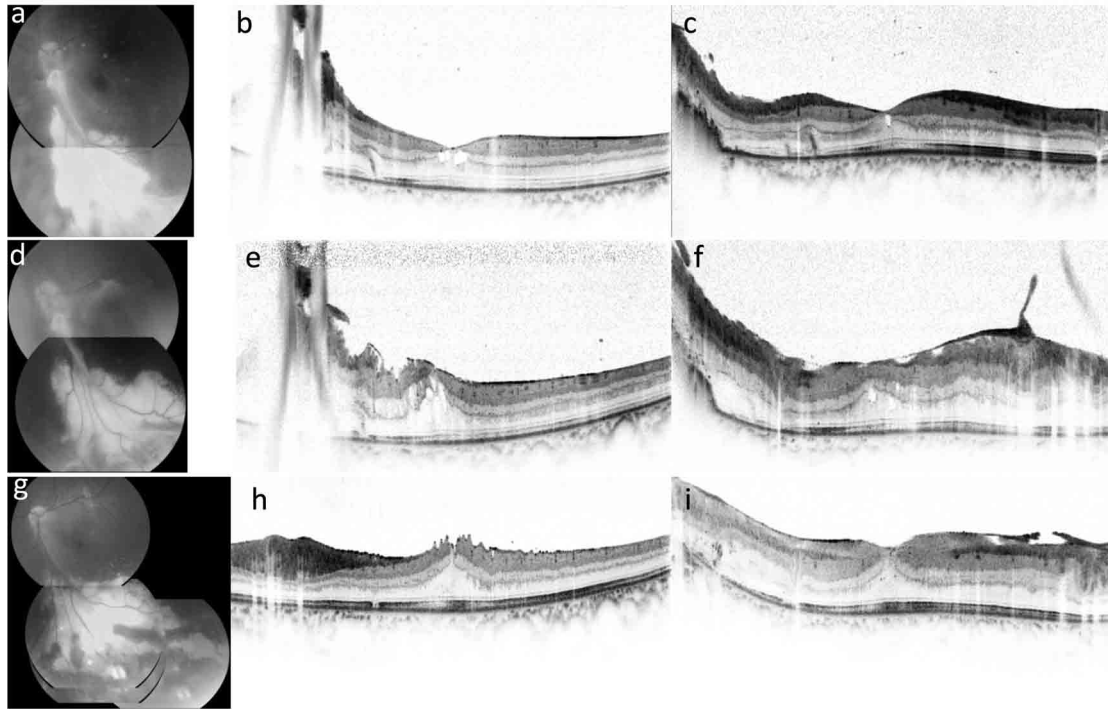


Figure 2. Fundus photographs and spectral-domain optical coherence tomographic (SD-OCT) images after treatment. (a, d, g) Fundus photographs. (b, e, h) SD-OCT images of a horizontal scan through the fovea. (c, f, i) SD-OCT images of a vertical scan through the fovea. There was no symptom of metamorphopsia or diplopia before surgery.

a-c : Four months after the initial visit, SD-OCT shows stronger traction on the disc margin and slight cystic changes after treatment with intravitreal bevacizumab injections, laser photocoagulations, and cryoretinopexy. Inferior dislocation of macula in addition to a flattened foveal contour and cystic macular edema suggests possible macular involvement. However, the decimal best-corrected visual acuity (BCVA) remained 0.7.

d-f : Nine months after the initial visit, traction to the parafoveal area is present causing moderate cystoid macular edema, and the BCVA decreased to 0.3. There is an epiretinal membrane (ERM) superior to the fovea, which was not seen 4 months after the initial visit (a), suggesting that the ERM might progress after intravitreal bevacizumab, laser photocoagulation and cryoretinopexy.

g-i : Thirteen months after 25-gauge pars plana vitrectomy, the traction on the parafoveal area is released and the BCVA has improved to 0.6. Macular edema and fine folds of the retinal surface remain, and a small regrowth of the ERM is seen at the superior arcade vessel.

DISCUSSION

Smithen *et al.* (11) reported that 7 of 13 patients (53.5%) with adult-onset Coats' disease had a final visual acuity of 0.3 or worse, and that 5 of 13 patients (38.5%) had visual acuity of 0.1 or worse. Although an ERM was noted in only 2.5 to 4.4% of eyes with Coats' disease in two earlier studies (5, 14), it is one of the major causes of visual acuity reduction in older patients. Smithen *et al.* (11) reported that an ERM was present in 2 of their 13 cases with adult-onset Coats' disease diagnosed after age 35 years (mean age of the 13 patients was 50 years). We suggest that Coats' disease complicated by an ERM is even rarer if the patient is in the 20's as in our case.

Inoue *et al.* (17) performed vitrectomy on 4 patients with juvenile Coats' disease with exudative detachment, and none had a complete PVD preoperatively. Wolfensberger *et al.* (3) reported that an ERM usually developed after a PVD had developed in the majority of patients with adult-onset Coats' disease. In our case with adult-onset Coats' disease, a PVD was detected at the initial visit.

The pathogenesis of the ERM in Coats' disease has not been conclusively determined, but Machemer (18) suggested that a chronic leakage from diseased vessels leads to reactive glial proliferation on the retinal surface. The development of ERM then causes traction on the retina and further accumulation of subretinal fluid.

To date, only two cases of untreated juvenile (3) or adult-onset (16) Coats' disease that underwent vitrectomy for an ERM have been reported. Wolfensberger *et al.* (3) reported a case of juvenile

Coats' disease with an ERM which continued to contract after laser photocoagulation. Then, vitrectomy led to an improvement of the BCVA from 0.1 to 0.5. Shukla *et al.* (16) reported that vitrectomy led to excellent anatomical and functional outcomes in adult-onset Coats' disease with an ERM if vitrectomy was performed before macular exudation and subretinal fibrosis developed. In their cases, the BCVA improved from 0.17 to 1.0 after vitrectomy with laser photocoagulation and peeling of the ERM and ILM. They emphasized that early vitrectomy and simultaneous endophotocoagulation is necessary for Coats' disease with an ERM before submacular exudation and subsequent submacular fibrosis developed.

In our case, the traction on the parafoveal area was released and the BCVA improved to 0.6 after the vitrectomy. However, the macular edema and fine folds of the retinal surface remained, and the BCVA did not change during the 13 months follow-up period. There is a possibility of better visual outcome, if vitrectomy had been performed 4 months after the initial visit when an increase in the traction on the disc margin was first detected.

On the other hand, Lafaut *et al.* (19) and Sugimoto *et al.* (10) reported cases of Coats' disease with spontaneous peeling of the ERM after laser photocoagulation. Both groups suggested that the PVD induced by laser photocoagulation led to the peeling of the ERM which was attached firmly to the posterior vitreous membrane. In fact, Sugimoto *et al.* (10) observed that a PVD developed after laser photocoagulation, and that the ERM detached from the vitreous concomitantly with the development of the PVD. Because the ERM was too firmly attached and wide spread in our case, a spontaneous peeling would not be expected.

Vascular endothelial growth factor (VEGF) has been identified as a key regulator of angiogenesis and vascular permeability (20). Lin *et al.* (20) reported that the mean intraocular concentration of VEGF in 4 eyes with Coats' disease was 2394.5 pg/ml which was significantly higher than the 15.3 pg/ml in 5 eyes with rhegmatogenous retinal detachment. There have been several reports on the effect of anti-VEGF therapy with intravitreal bevacizumab alone or in combined with laser photocoagulation in Coats' disease. These treatments benefited Coats' disease cases with macular edema and exudative retinal detachment (21-23). However, the use of anti-VEGF therapy has not been proven to have marked effect on the visual outcomes (24). For adult-onset Coats' disease, Ramasubramanian *et al.* (22) reported complications such as vitreo-retinal fibrosis and traction retinal detachment after intravitreal bevacizumab combined with standard therapy. Similarly, traction by the ERM was increased after intravitreal bevacizumab, laser photocoagulations, and cryoretinopexy in our case.

Shienbaum *et al.* (2) reported on the recurrent nature of Coats' disease. They followed 12 patients treated for Coats' disease for an average follow-up period of 12.4 years. Four of the 12 patients (33%) had recurrences, and three of the four had multiple recurrences. The average elapsed time from successful treatment to the first recurrence was 4.3 years, and the average number of recurrences was 3.3. Shukla *et al.* (16) raised caution about late recurrences of ERMs many months or years after a successful vitrectomy for ERM secondary to Coats' disease. Consistent with their findings, a small ERM regrowth developed in the superior arcade vessel 13 months after vitrectomy in our case. No intraoperative ILM peeling might contribute to this ERM regrowth.

In conclusion, 25GPPV combined with ERM peeling, laser photocoagulation, and cryoretinopexy may be an effective treatment regimen for adult-onset Coats' disease with an ERM. However, the patients should be examined periodically to detect recurrences.

CONFLICT OR COMMERCIAL INTEREST

None for each author

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